THE SURGICAL TREATMENT OF TUMORS OF THE PERIPHERAL NERVES

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THE multiplicity in types of tumors of the peripheral nervous system and the diversity with which these lesions are found in the face, neck, extremities, thorax and abdomen, have given rise to an extensive literature on descriptions of such growths and their proper surgical treatment. The different forms of these neoplasms occurring in widely scattered parts of the body call for operative procedures of great variety, and often tax the surgeon's ingenuity in treating them. There are, however, certain general principles which can be set forth to guide the treatment of nerve tumors as they are attacked at various sites and in the following presentation we briefly review some of the accepted ideas on technical procedures, choice of operations, and postoperative care. We have purposely avoided discussion of the nerve sheath tumors arising in the abdomen, principally the stomach, for the problems of removal are essentially the same as those for other intra-abdominal neoplasms. The nerve sheath tumors of the mediastinum and thoracic cage necessitate a detailed and rather specialized technic which is beyond the scope of the present communication, but valuable contributions have been made to this field, particularly by Harrington[10] and Heuer.[11] In the following discussion* some of the more commonly employed procedures are described first, leaving until later other considerations such as selection of operation and postoperative care.

Preoperative Considerations.—The anesthetic employed need give little concern, for a wide variety can be chosen from if there are no complicating factors. Ether, avertin-ether, or nitrous oxide-oxygen-ether all serve admirably. Spinal anesthesia is preferable for operations upon the legs. One should not attempt, however, to remove one of these neoplasms under local conduction anesthesia, for injection of novocain directly into a large nerve trunk is too apt to produce injury and subsequent neuritis.

The armamentarium need include but few special instruments. A blunt end nerve hook will be found desirable in handling small trunks. Delicate smooth and mouse tooth forceps are essential. Silver clips may be found useful for controlling bleeding vessels on the surface of or in the substance of a nerve. Fine silk is far superior to catgut if repair or end-to-end suture of a nerve trunk is to be done. Black silk of size 1 is suitable, but if this is not available, No. 4 (A silk) can be split into its three separate strands

* Three of the cases cited here for illustrative purposes were operated upon by Dr. Harvey Cushing, Dr. David Cheever and Dr. John Homans to whom we make grateful acknowledgment.

and one of these will be of correct diameter and strength. Commercially prepared blood vessel suture silk is excellent. The needles should be small, curved, semi-curved and straight, with closed eyes. These can be threaded beforehand and if sterilized in oil or waxed they will be found to slip more easily through nerve tissues if repair is performed.

Operative Procedures.—*Excision of Tumor from Within Trunk Without Division of Functionally Active Fibers.*—If the tumor is a benign neurofibroma or neurilemmoma, it is permissible to shell it away from the nerve trunk. It is of prime importance to recall that the benign nerve sheath tumor (unassociated with von Recklinghausen's disease) grows in such a way as

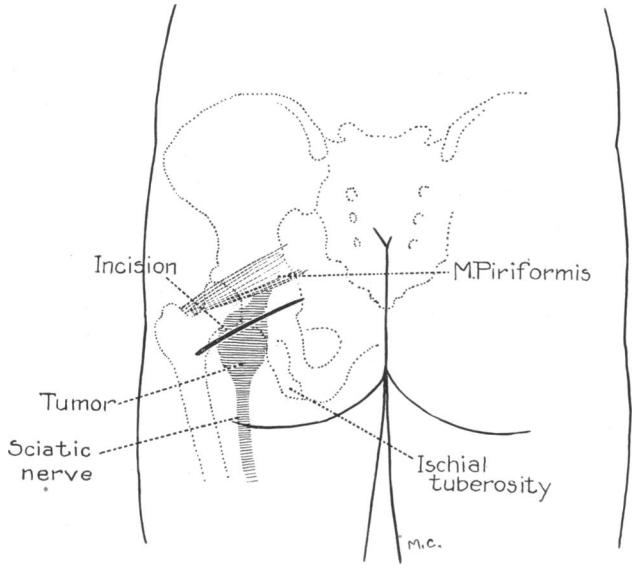

Fig. 1.—Schematic drawing to show position of a benign peri neurial fibroblastoma of the left sciatic nerve from a case treated as indicated in Figs. 2, 3 and 4.

to push the nerve trunk to one side or to expand it. The nerve proper does not, therefore, directly enter the mass but is displaced laterally or is pushed out so as to completely surround the neoplasm. Thus one always finds a good line of cleavage which permits dissection of the tumor away from the nerve without severing its fibers or disturbing their functional integrity. When the nerve lies toward one side, it can, of course, be dissected off with ease. If, however, the nerve completely surrounds the mass, the covering fibers must be incised longitudinally to permit withdrawal of the central growth. Figs. 1 to 4 illustrate such a treatment in removal of a lemon-sized perineurial fibroblastoma of the sciatic nerve in one of our cases. After this dissection the nerve may be frayed out or indeed may consist of little more than a flabby, collapsed shell, but it can be dropped back into the wound and there may be little or no sensory or motor disturbance in the area which it supplies. If longitudinal incision has been made in the nerve trunk to permit

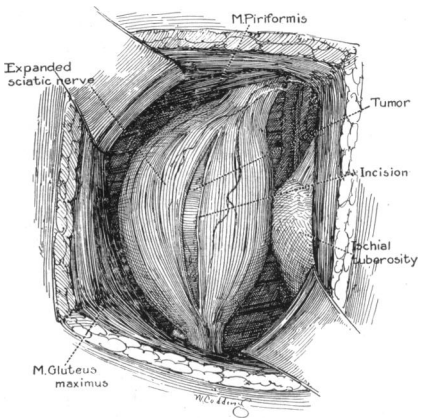

FIG. 2.—Operative exposure of a perineurial fibroblastoma of the sciatic nerve in the buttock (compare with Fig. 1). The gluteus maximus muscle has been split and the edges retracted. Fibers of the sciatic nerve are expanded around the neoplasm. Incision is made parallel to the nerve fibers in order to reach the central mass.

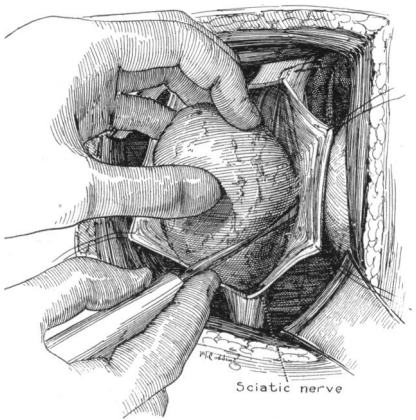

FIG. 3.—Removal of a benign nerve tumor from the sciatic nerve (see Figs. 1 and 2). The sciatic nerve is split open and its edges gently retracted with silk stitches. The neoplasm is shelled out from its bed by blunt dissection.

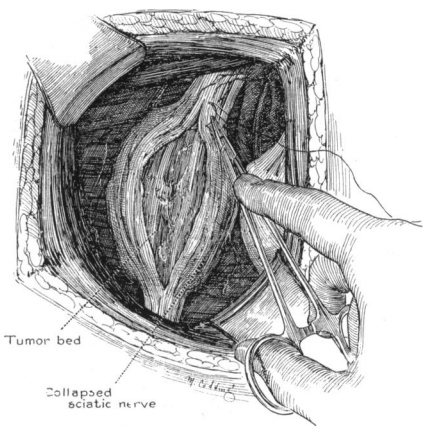

FIG. 4.—Repair of nerve trunk following extirpation of a neurofibroma (see Figs. 1 to 3). The separated edges of the nerve are lightly approximated by a few silk stitches. The nerve, though stretched and flabby, remains functional.

enucleation of a tumor, the edges of this defect can now be brought together with a few interrupted silk sutures (Fig. 4). Such approximating sutures should be of the finest silk and loosely tied so as not to injure apparently healthy nerve fibers.

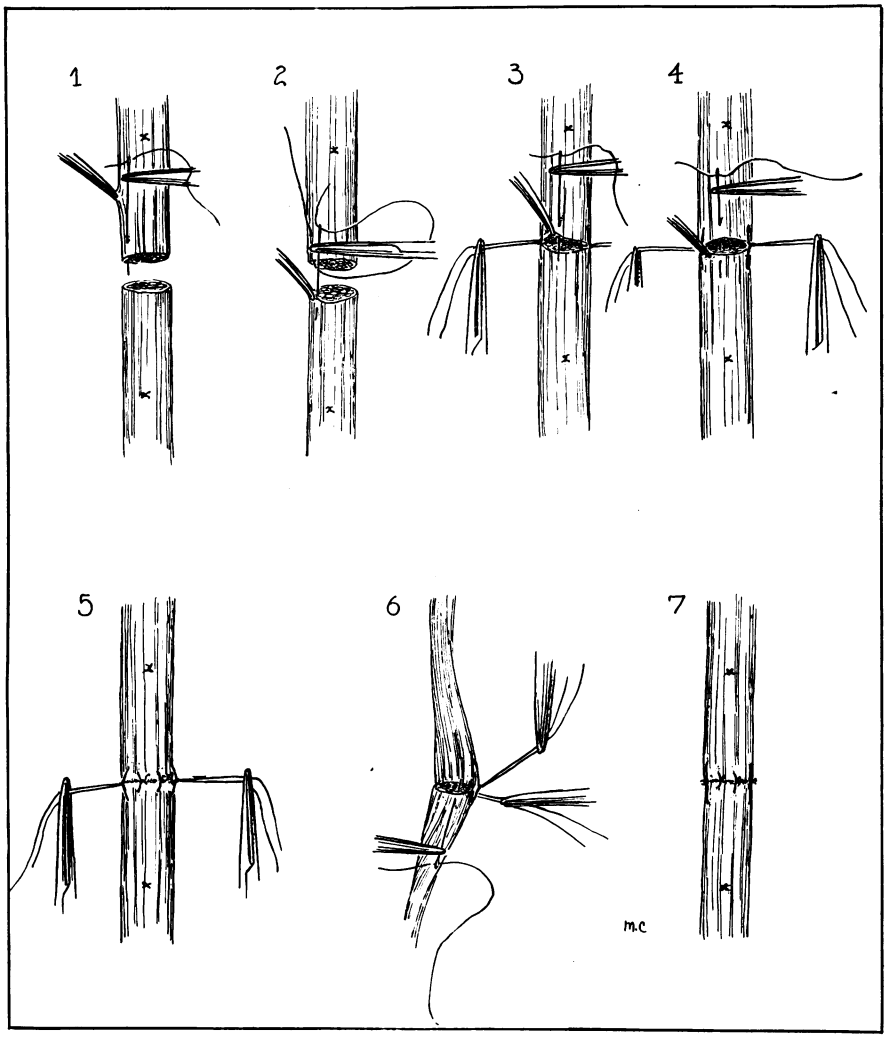

FIG. 5.—Method of end-to-end nerve suture. Note in all of the sketches the small identification knots (placed before the nerve was divided) which serve to prevent rotation of the proximal or distal segments. (1) Fine straight needle passed through epineurium. (2) Stitch carried through covering of opposing nerve end. (3 and 4) First two (diametrically opposite) stitches left long and snapped for purposes of handling the nerve. Intermediate sutures being placed. (5) Sutures completed on front of nerve. (6) Nerve rotated for placing of posterior sutures. (7) Repair completed.

Excision of Tumor with Division of Nerve.—The second and less commonly employed method of nerve tumor excision is to make a clean and complete resection of the mass so as to include a portion of nerve proper. If the nerve which is thus surgically severed is a small subcutaneous one or is

an unimportant branch, no attempt at repair is necessary. If, however, the trunk is large, it is definitely the duty of the surgeon to attempt an immediate restoration by end-to-end sutures. Such a reconstruction is relatively easy to perform, but care and delicacy must be exercised to establish a nice approximation of the divided nerve ends in order to reduce the possibility of subsequent neuroma formation and to give the best possible chance for reestablishment of nerve function.

A number of methods have been described for end-to-end nerve suture and the steps indicated in Fig. 5 represent an acceptable operation. The ends of the nerve must be cut cleanly at right angles to the long axis of the trunk. Two guy sutures are then placed (Fig. 5). These sutures include 2 or 3 Mm. of the nerve end and pass through the ensheathing epineurial coat only, for this is the strongest portion of the nerve and must be employed to give

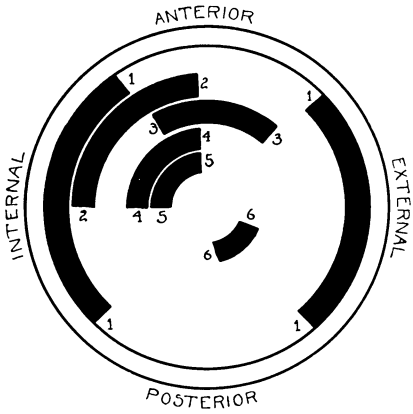

FIG. 6.—Schematic cross-section of median nerve at the midarm level (from Kraus and Ingham) showing position of bundles as determined by electrical stimulation of this level. (1) Pronator radii teres; (2) flexor carpi radialis; (3) palmaris longus; (4) flexores digitorum; (5) flexor pollicis; (6) pronator quadratus.

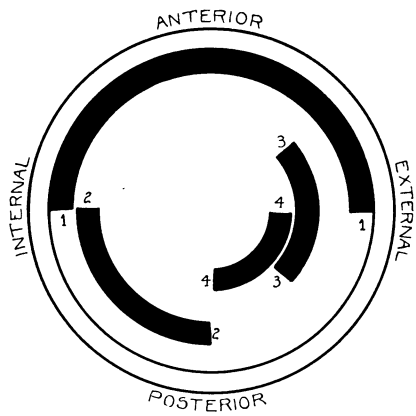

FIG. 7.—Schematic representation of funicular topography of median nerve at midforearm level (from Kraus and Ingham). (1) Sensory; (2) opponens pollicis; (3) abductor pollicis; (4) lumbricales.

strength to the suture line. Under no circumstances should these sutures pass diametrically through the nerve, for in such a location they tend to buckle the nerve union and to impede the progress of regenerating axis cylinders. These first two stitches are left long and are used for traction in order that the nerve may be held and rotated during placement of the remaining sutures (steps 5 and 6, Fig. 5). Babcock[1] has devised a clamp by means of which the two ends of the nerve may be held by small piercing wires and thus be brought together and handled during the steps of suturing. With the ends of the nerve drawn together, additional interrupted sutures may now be placed around the entire periphery of the nerve, the total number varying from four or five in a small trunk to as many as 12 or 15 in a large nerve such as the sciatic. The nerve should be picked up with the finest of forceps, and only the outer covering need be grasped during the manipulations. The

nerve tissue tends to dry quickly and hence should be kept moist with saline while it is exposed.

When any nerve is divided and repaired by end-to-end suture, great care must be taken to prevent rotation of the two cut ends when they are sutured. It has been shown that fibers coursing to muscle groups are isolated into more or less segregated portions of a nerve trunk (Figs. 6 and 7), and hence

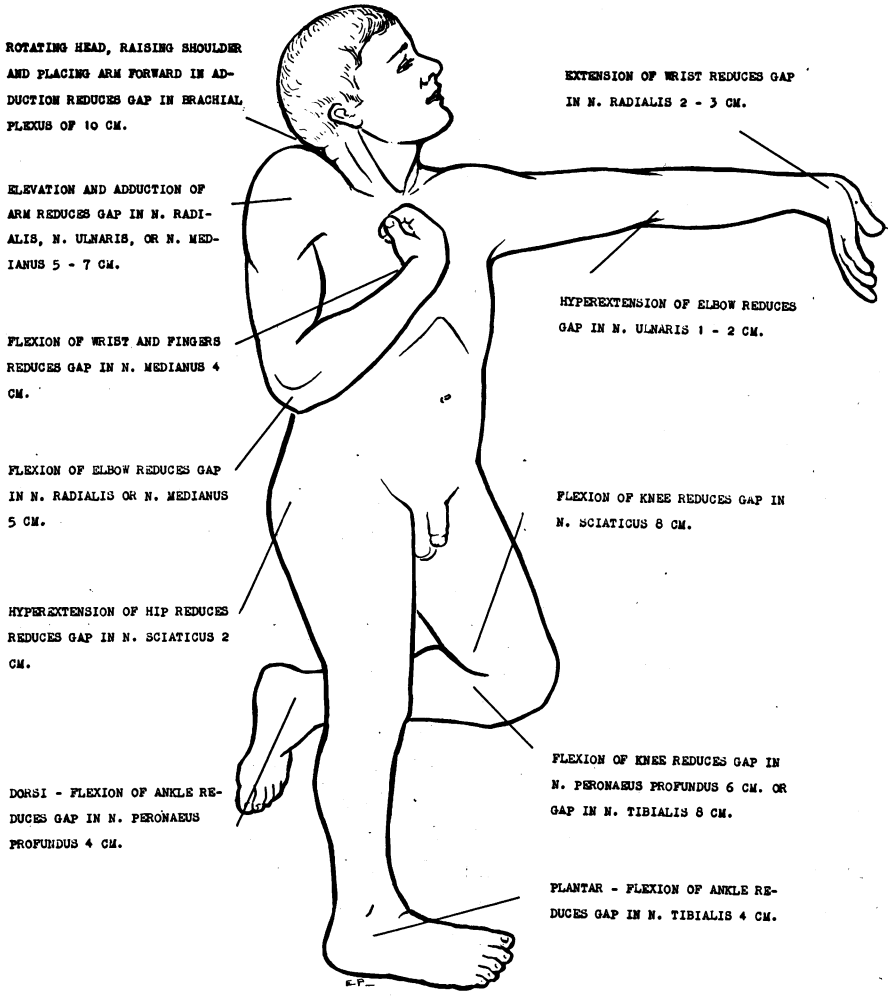

FIG. 8.—Diagrammatic representation of the amount of nerve defect which may be overcome by movement of joints as indicated (after Babcock).

an attempt must be made to join the two ends so that the bundles of the proximal portion will lie in apposition to the corresponding funiculi of the distal end. This will insure the best possible chance for the regenerating axones to grow out into their proper peripheral distribution. A good way to guard against rotation of the two ends is to place two small silk ties on

one surface of the nerve before excision of the tumor. These knots then serve as identification sutures during the various stages of end-to-end suture (Fig. 5).

The great size attained by many of the peripheral nerve tumors may appear to militate against excision of the nerve trunk for fear of not being able to subsequently bring the divided ends together. The general rule may be stated, however, that one never encounters a nerve tumor so large that removal of the involved nerve trunk prohibits bringing together of the divided nerve ends. The only exceptions to this are provided by neoplasms of the thorax and, rarely, by some of those arising in the neck. In the arms and legs, however, it is remarkable what lengths of nerve gaps may be closed by employing one or several of the following important procedures:

(1) The normal elasticity of the nerve allows it to be stretched for lengths varying from 1 to 2 cm., depending upon the nerve under consideration. This small but important degree of slack may be utilized without placing undue tension on the suture line.

(2) The proper position of a limb adds a considerable additional available length which may be employed in reducing a nerve gap. Babcock[2] has had a most extensive experience with peripheral nerve injuries and has employed this method with unusual success, reducing the nerve gaps in various nerves by the maximum amounts indicated in Figure 8.

(3) Additional lengths may be obtained in a nerve trunk by rerouting. Not all nerves lend themselves to this procedure, but a familiarity with the technic will occasionally permit a reduction of a nerve gap not obtainable by any other operation alone. The ulnar, median, radial and tibial nerves offer the best branches for this procedure. The ulnar nerve may be loosened from its bed at the elbow and by changing its position so as to make it lie in front of the internal epicondyle of the humerus, an additional 2 cm. may be gained. The radial nerve may be unwrapped from the humerus and upper arm muscles, and by displacing it to lie on the anteromedial aspect of the upper arm an additional 3 to 4 cm. may be added to its length. The median nerve can be displaced from its deep bed and by raising it to a position superficial to the forearm muscles a length of 2 cm. can be gained. In performing these procedures great attention must be paid to the twigs supplying surrounding muscles and these individual fibers may be stripped back along the nerve so that they take origin higher up than they did formerly. By this procedure these muscle branches are damaged only a negligible amount. The tibial nerve can be displaced to a position in front of the internal malleolus, an additional 2 to 3 cm. being gained thereby. Rerouting of a nerve necessitates very liberal exposure and unless one is prepared to make an unusually long incision, the operation is doomed to failure.

Table I indicates the maximum amount of gap which may be reduced in various nerves (Babcock) by taking advantage of the normal nerve elasticity by proper position of the neighboring joints, and by rerouting.

TABLE I

MAXIMUM GAPS IN PERIPHERAL NERVES IN WHICH END-TO-END SUTURE IS POSSIBLE (AFTER BABCOCK)

	By Slack and Elasticity	By Joint Position	By Rerouting	Totals
	cm.	cm.	cm.	cm.
Brachial plexus	1.5	3–7	....	11.5
Radial in arm	3.0	5–7	....	15.0
Radial in forearm	1.5	4–5	....	10.5
Ulnar in arm	3.0	7	6	16.0
Ulnar in forearm	1.5	5	6	12.5
Median in arm	3.0	5–7	....	15.0
Median in forearm	1.5	3–4	14.5	23.0
Sciatic	3.0	3–8	....	14.0

The plexiform neuromata of von Recklinghausen's disease are so variable as to make it impossible to standardize the different therapeutic procedures. Occasionally, however, one is fortunate enough to encounter a localized form of the disturbance and the irregular, nodular enlargements of the superficial or subcutaneous nerves can be easily dissected out. As the twigs involved are often small or sensory ones, no nerve repair is usually required. If the overlying skin is pigmented or pedunculated, portions of it may be excised to gain a good cosmetic result, the cutaneous defect being closed by plastic procedures as the circumstances demand.

The neck is one of the most common sites for the nerve sheath tumors, particularly the benign perineurial fibroblastoma, which may arise from any one of the numerous sympathetic, cranial or somatic nerves which richly traverse this region (Figs. 9 and 10). One must be prepared to dissect deeply for these neoplasms which may be attached to the roots of the cervical plexus or extend to the very base of the skull if they arise from the vagus. When they originate from the cervical roots (most commonly the dorsal) dumb-bell shaped or hour-glass tumors[11] may lie partly in the spinal canal and also out in the soft tissues of the neck, hence the unwary operator may be led into a more extensive undertaking than had been anticipated. If the nerve trunk is divided in removing one of these cervical neoplasms, every effort should be made to resuture the cut ends, this consideration being most important in division of the hypoglossal nerve in order to prevent extensive atrophy of the tongue muscles.[7] On two occasions we have purposely divided the left vagus nerve without being able to repair it, and in neither case could we detect any disturbance in the cardiac or intestinal mechanisms post-operatively. In one of these cases a permanent hoarseness followed operation. On the whole, however, one commonly finds that it is possible to peel these cervical tumors away from the nerves which give rise to them and it is rarely necessary to completely divide one of these important trunks.

The thorax, while presenting relatively few of the nerve sheath tumors,

contributes an especially interesting chapter for the benignity of these neoplasms in the mediastinum or chest wall and makes their removal a most satisfactory undertaking. One of these cases in our series had a hen's egg size perineurial fibroblastoma of the superior mediastinum which was successfully removed by opening into the mediastinum through the sternum. The upper portion of the sternum was transected between the first and sec-

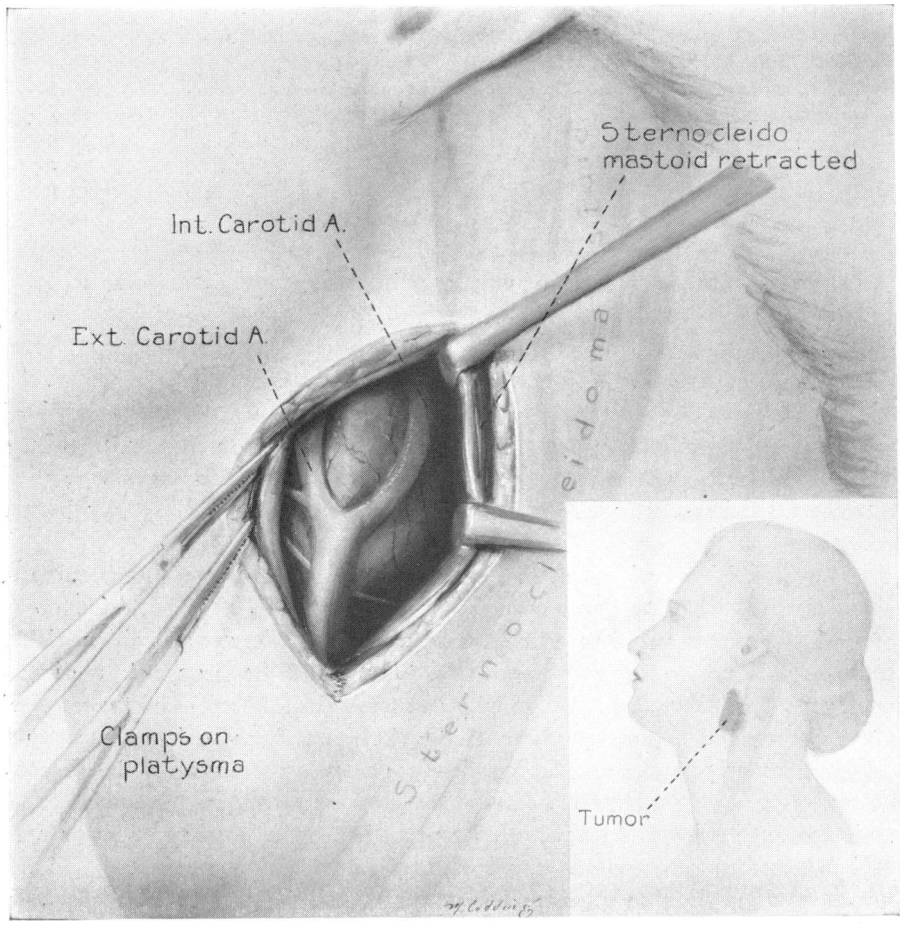

FIG. 9.—Operative exposure of a deeply attached cystic neurofibroma of the neck. A segment of the internal jugular vein has been resected to gain access to the neoplasm. The external carotid artery is compressed anteriorly and the internal carotid artery is pushed laterally. The mass was cleanly excised. Insert shows position of the tumor.

ond ribs and between the third and fourth ribs by the use of a Gigli saw. Cutting through the second and third costochondral junctions on either side then permitted the turning to the right of a skin, muscle, and bone flap, leading into the mediastinum as through a trap door (Fig. 11). Keller and Callender[14] removed a neurofibroma at the lower end of the left phrenic nerve, approaching it by an incision through the left fifth interspace, dividing and spreading the fifth and sixth ribs upward and downward respectively.

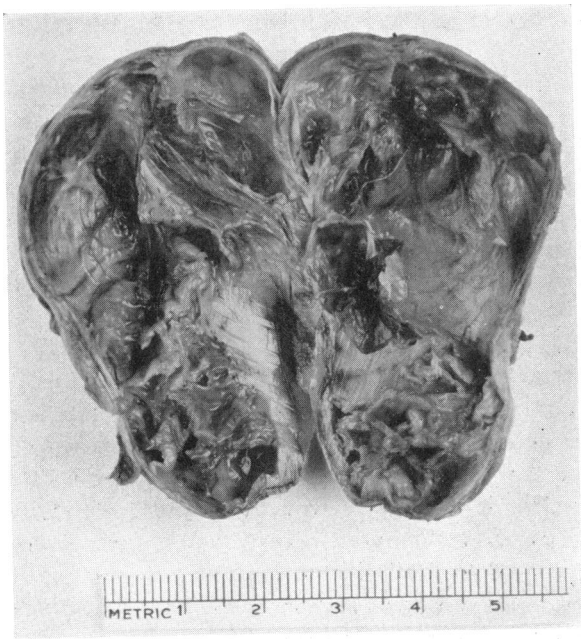

FIG. 10.—Photograph of neurofibroma of the neck showing extensive cystic degeneration. (From same patient as Fig. 9.)

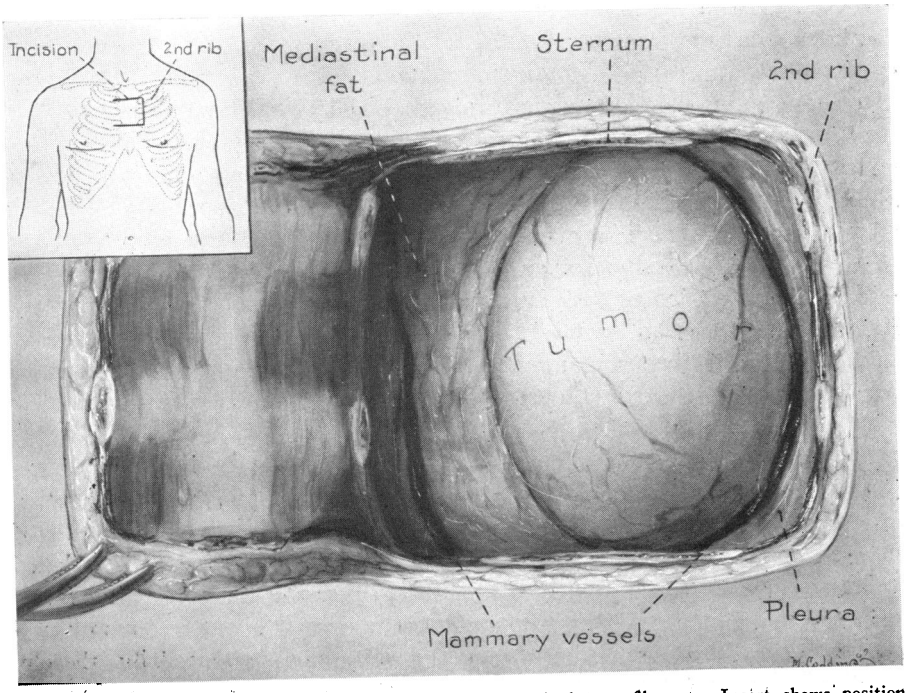

FIG. 11.—Sketch from successful removal of a mediastinal neurofibroma. Insert shows position of operative incision. A trapdoor, including skin, subcutaneous tissues and upper portion of sternum has been turned to the patient's right. The tumor was shelled out from its bed and dissected away from the parietal pleura on its left and the great vessels which lay deeply toward its right.

This gave good access to the region between the pericardium and the left parietal pleura.

In one of our patients with a large orange size perineurial fibroblastoma of the posterior portion of the right eighth intercostal nerve a successful and easy extirpation was completed by removing the posterior one-half of the ninth rib subperiosteally and then cutting across the eighth and tenth ribs posteriorly so that these latter could be spread apart. The pleural cavity was then entered by an incision through the bed of the ninth rib and the tumor, which projected into the thoracic cage, was easily excised from its attachments to the eighth intercostal nerve.

The posterior mediastinum is by far the best known site for the nerve tumors of the thorax. Since the spinal cord roots as well as the sympathetic nerves account for these posterior tumors, one finds that they lie close to the spinal column and extrapleurally. Thus they can usually be removed without entering the pleural cavity if a careful dissection is made. The operative approach then is usually by way of a paravertebral incision, cutting through the necks of two or more ribs to permit their separation. Again, one must be prepared to deal with an hour-glass tumor, opening the spinal canal at the same or subsequent sitting as the findings demand.

Postoperative Care.—Other than the usual care of the wound, little need be said concerning postoperative treatment excepting in those occasional cases in which a nerve trunk has been divided and resutured. When this latter has been done, the postoperative treatment should include an immobilization of the extremity, a graded return of motion of the part, and physiotherapy to those regions which are temporarily deprived of their nerve supply.

Immobilization is required to permit proper healing of the suture line and to give the best possible chance for the regenerating axis cylinders to grow down the distal part of the nerve trunk. It is especially necessary when a nerve gap has been overcome by flexing (or otherwise adjusting) the contiguous joints, because movement of these joints may tear apart the nerve repair. The duration of such immobilization must, of course, be decided for the individual case, but certainly it should never be less than three or four weeks and should better be maintained until there is evidence of return of nerve function. It is safer to err on the side of immobilizing too long rather than too short a period.

A graded return of motion should be ordered for the immobilized extremity. If a joint has been fixed in a given position to permit obliteration of a nerve gap, it is best to adopt some set schedule for reestablishment of the normal range of motion of this joint. Thus, if the elbow has been flexed at a 90° angle to permit suture of the median nerve in the antecubital fossa, it is best to maintain this right angle for a period of about one month and then to increase the extension at the elbow by two degrees daily, thereby passing a total of two and one-half months before getting the forearm straightened out to 180°.

Physiotherapy has a definite place in the postoperative regimen, for by

its judicial use the distal musculature and soft parts can be maintained in the best possible condition, and atrophy partially prevented while waiting for regeneration of the nerve. Baking and massage given daily or three times a week as the economic conditions warrant should be continued until restoration of active motion in all the paralyzed muscles heralds the return of nerve function. Physiotherapy can be administered while the limb is partially loosened but not removed from such immobilization apparatus as might be employed. Former teachings advocated the use of electrical stimulation of paralyzed muscles but experience has shown that this has little additional advantage and hence can be omitted with impunity.

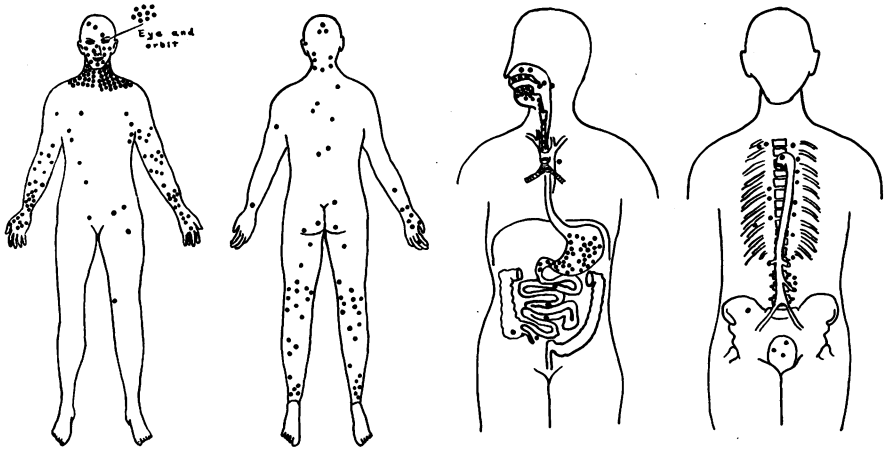

FIG. 12.—Distribution of benign perineurial fibroblastoma (from Stout, to which are added 17 cases of Mayo and Barber and 20 personally observed cases).

The length of time necessary for reestablishment of muscular and sensory function, naturally, depends on the length of the nerve trunk distal to the point of its severance and end-to-end suture. In general, under optimum conditions, the axis cylinders will regenerate at the rate of about 1 Mm. per day or roughly one inch per month. If there is no evidence of returning nerve function in twice the time calculated and allotted for its reestablishment, it is likely that the nerve suture has broken down and the ends separated or else a neuroma has formed at the site of operation and secondary suture in all probability will be required.

Choice of Treatment in Various Types of Peripheral Nerve Tumors.—The important tumors of the peripheral and sympathetic nervous systems may be classified as follows:

(1) von Recklinghausen's disease (neurofibroma).
(2) Perineurial fibroblastoma (neurilemmoma, schwammoma).
(3) Malignant perineurial fibroblastoma (malignant neurofibroma).
(4) Neurofibrosarcoma (neurogenic sarcoma).
    Arising in von Recklinghausen's disease.
    Arising in perineurial fibroblastoma.

(5) Cyst (?degenerated perineurial fibroblastoma).
(6) Neuro-epithelioma.
(7) Ganglioneuroma.

von Recklinghausen's disease in its generalized form usually does not call for operative treatment, but when it occurs as an isolated lesion or in the "forme frustes" the possibility of complete eradication makes surgical intervention advisable. Such plexiform neuromata can be dissected out with more or less ease, depending upon the extent and depth of the pathologic process. The generalized types of the disease may require surgery when large pedunculated masses cause discomfort and when undue rapidity of

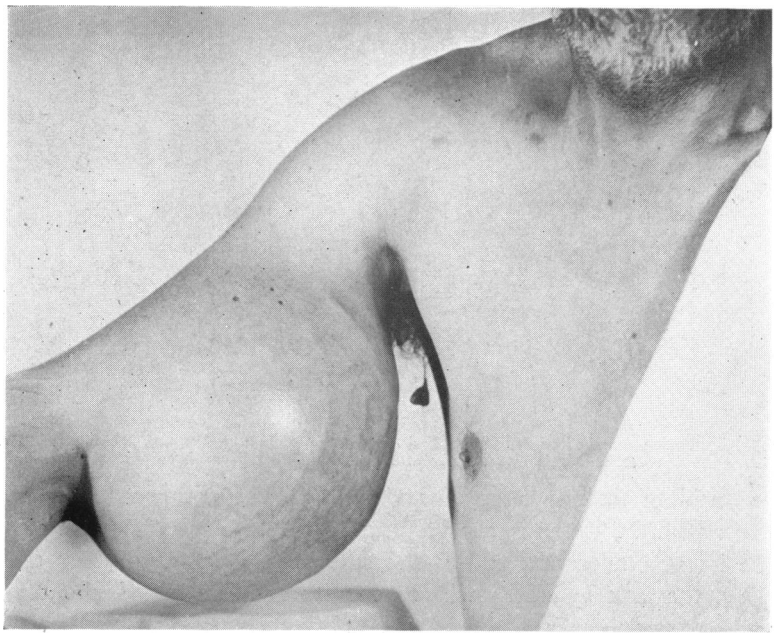

FIG. 13.—Neurofibrosarcoma of the median nerve in a 56 year old man. Small mass had been present for 20 years—followed by recent rapid growth and doubling of size in four months. Apparently malignant change in a previously existing neurofibroma. (No stigmata of von Recklinghausen's disease.)

growth suggests the possibility of malignant degeneration—a transformation which occurs in about 15 per cent of cases of von Recklinghausen's disease.

The perineurial fibroblastoma can practically always be treated by conservative surgery, for these lesions are slowly growing and if cleanly removed they rarely recur. When attached to small subcutaneous nerves, removal of a bit of the nerve causes little disability, but if the mass arises from a large nerve, it should be peeled away so as to leave the trunk intact. Figure 12 indicates the common positions of these growths, and as is seen, the most frequently involved sites are the neck, the front of the arms, and the backs of the legs.

The term malignant neurofibroma has been adopted by us to designate

several specimens which have shown a marked tendency to local recurrence and regional extensions. These uncommon growths do not exhibit a high degree of malignancy and do not metastasize by way of blood stream or lymphatics. They do, however, produce important local and regional invasions. While some authors have classified these neoplasms as slowly growing neurofibrosarcomata, we have preferred to employ a different term in speaking of them in order to point out their activity, which lies halfway between the neurofibroma or perineurial fibroblastoma on the one hand and the neurogenic sarcoma on the other. Their surgical treatment calls for a local but radical resection, and under no circumstances should attempts be made to separate them from the nerve trunk for this will almost certainly lead to recurrence and will doom to disaster a condition which previously had been

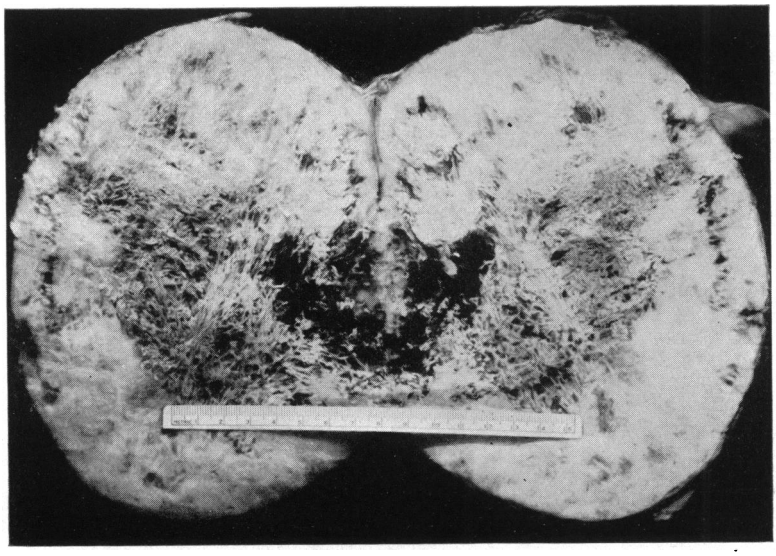

FIG. 14.—Rapidly growing neurofibrosarcoma of the median nerve. Specimen from case illustrated by Fig. 13.

curable. The excision, therefore, should always include a portion of the nerve trunk, which can be repaired at the same sitting by an end-to-end suture. We have justified this procedure in our own experience by removing such a neoplasm with a segment of the posterior cord of the brachial plexus. In spite of the postoperative total paralysis of radial and axillary nerves, the muscular and sensory functions of these nerves were completely restored in the arm within two years and there has been no evidence of recurrence of the neoplasm.

The neurofibrosarcomata, in the absence of demonstrable metastases, should be attacked radically from the outset. No form of local removal should be considered adequate if the growth appears on a limb, for early amputation may be a life-saving procedure. If pathologic examination of a specimen shows a sarcomatous growth in what was preoperatively thought to

be a benign lesion, amputation can be performed secondarily with some hope of cure. It must be noted, however, that one's suspicions concerning malignancy should be aroused before operation for there is almost invariably a history of rapid growth in a period of a few months or else there has been a sudden increase in size in a previously existing long standing benign tumor. If, therefore, a recent rapid growth has been noted, preparations should be made for competent pathologic examination at the time of operation, so that amputation can be done immediately if necessary. It is almost safe to say, however, that if there has been a recent marked increase in the size of the mass and if at operation there is no hemorrhage or cystic degeneration to account for this, the tumor may be considered almost certainly a malignant one. Figs. 13 and 14 are from a case of neurofibrosarcoma of the median nerve in a man 56 years of age. Fig. 15 shows the distribution of 137 malignant, nerve sheath tumors as compiled by Stout.

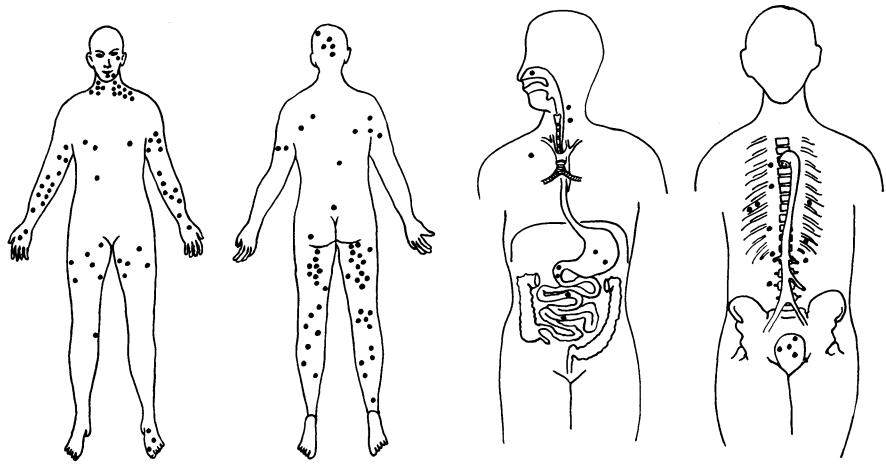

FIG. 15.—Distribution of 137 malignant nerve sheath tumors, occurring in 100 patients with von Recklinghausen's disease and in 29 patients without such stigmata (from Stout).

Cysts of the nerve trunk occur with rarity and their origin is not clearly understood. Since it is well known, however, that marked cystic degeneration often takes place in a perineurial fibroblastoma, it is only reasonable to believe that these nerve cysts, filled with watery or gelatinous fluid, take their origin from a previously existing nerve sheath tumor. The treatment, therefore, should be conservative, and consists of a simple dissection of the thin-walled sac away from the adjacent or surrounding nerve with as little interruption of the nerve fibers as is possible. The nerve can be split longitudinally so as to expose the cyst which lies within its substance, and by this technic the functional integrity of the nerve is not impaired.

Treatment of nerve cysts raises another possibility, namely, aspiration of the cyst or aspiration combined with injection of sclerosing fluids. Such therapy need only be rarely employed, but on one occasion we found it useful in treating a cystic, malignant neurofibroma of the face. In this patient a

large external mass in the region of the left zygomatic bone, extended internally to displace and involve the left side of the soft palate. Complete surgical removal was obviously impossible because of the anatomic location and only enough tissue for biopsy purposes was removed. Aspiration of the cyst, however, afforded considerable relief to the patient and the injection of fixative agents theoretically killed off a narrow rim of tumor remaining around the cyst wall.

The neuro-epitheliomata are lesions of an extraordinary high degree of malignancy and the same may be said regarding their treatment as was noted regarding the neurofibrosarcomata.

The ganglioneuromata are tumors of not too common occurrence, but their relative benignity and the fact that more than one-third of them are seen in childhood makes surgical removal a satisfactory and worthwhile procedure. McFarland and Sappington[19] collected 143 examples of this tumor from the literature. Among these cases the three most commonly involved sites were: retroperitoneal, 26; cervical, 17; thorax and mediastinum, 11. These neoplasms are well encapsulated, usually quite slowly growing and can be removed by local excision with a considerable degree of success and freedom from recurrence.

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